

into the small intestine. A thin tube is placed through the scope into the bile ducts, and dye is injected to highlight the bile ducts on the x-ray.

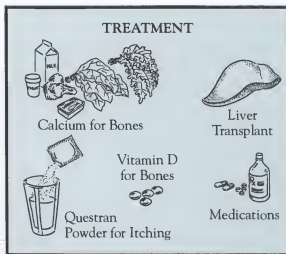
As the disease progresses, a liver biopsy is needed to determine how much damage has occurred. Under local anesthesia, a slender needle is inserted through the right lower chest to extract a small piece of liver for microscopic analysis.

Treatment

Because PBC advances slowly, patients often have no symptoms for many years. Initial treatment for PBC is aimed at reducing symptoms when they occur. Itching can be controlled by drugs such as Questran. Bile is usually reabsorbed into the bloodstream from the large intestine, and goes back to the liver to be reused. Questran binds up bile in the intestine, allowing it to be eliminated with the stool instead. This helps to reduce the build-up of bile in the body.

The diet should be well-balanced. At least 1,200 mg of calcium per day is needed to prevent osteoporosis. Also, if blood levels of vitamin D are below normal, the physician may prescribe a supplement. Vitamin D helps the body absorb calcium from the intestine. If thyroid function is low, it too can be treated with medication. Diuretics (fluid pills) and reducing salt intake can help reduce edema or swelling.

Some drugs seem to improve liver function blood tests. Some research studies suggest these drugs reduce damage to the liver. Actigall changes the make-up of bile in the liver and seems to reduce liver damage. Colchicine is an older drug that may reduce scar formation in the liver. Another drug that shows promise is methotrexate (trade name Rheumatrex). It suppresses the immune response in the body. When PBC progresses to a point where too much liver damage has occurred, liver transplantation must be considered.



Liver Transplantation

Liver transplantation is now an accepted form of treatment for chronic, severe liver disease. Advances in surgical techniques and the use of new drugs to suppress rejection have improved the success rate of transplantation. The outcome for PBC patients is excellent. Because of the disease's slow progress, it is possible to plan elective transplant surgery. Survival rates at transplant centers are well over 90 percent, with a good quality of life after recovery.

Summary

Primary biliary cirrhosis is a slow, progressive disease. Once diagnosed, treatment is directed at managing symptoms and slowing down liver damage. A great deal of research is underway aimed at discovering the cause, preventing damage to the bile ducts and liver, improving symptoms, and prolonging life. Transplantation is now a standard form of treatment for advanced disease. By working closely with the physician, there is good reason to expect a favorable long-term outlook.

This material does not cover all information and is not intended as a substitute for professional medical care.

PRIMARY BILIARY CIRRHOSIS

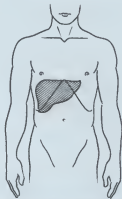
The Liver

The liver is the largest organ in the body. It is found high in the right upper abdomen, behind the ribs. It is a very complex organ and has many functions. They include:

- Storing energy in the form of sugar (glucose)
- Storing vitamins, iron, and other minerals
- Making proteins, including blood clotting factors, to keep the body healthy and help it grow
- Processing worn out red blood cells
- Making bile which is needed for food digestion
- Metabolizing or breaking down many medications and alcohol
- Killing germs that enter the body through the intestine

The liver cells excrete bile into tiny tubes within the liver called bile ducts. These tubes come together like the tiny veins on a leaf. They drain the bile into the common bile duct, a larger single tube leading into the intestine. There the bile aids digestion and gives stool its brown color. As you can see, the liver is a very important organ.

LOCATION OF LIVER



What is Primary Biliary Cirrhosis (PBC)?

Primary biliary cirrhosis is a disease of the bile ducts inside the liver. It progresses slowly, so patients may lead active, productive lives for many years. In PBC, the bile ducts in the liver become inflamed. The inflammation is chronic (constant over a long period of time), and causes scarring that eventually blocks and destroys the bile ducts. This condition interferes with the proper drainage of bile, so the bile backs up into the liver and into the bloodstream, causing various symptoms. Eventually the liver itself becomes badly damaged and scarred. This is known as cirrhosis.

Cause

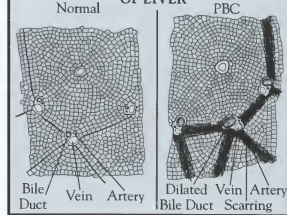
The exact cause of PBC is unknown. Scientists believe there could be more than one contributing factor. While it does not have the traits of an inherited disease, it does appear more often in some families. People with PBC sometimes have a history of allergies or autoimmune disturbances—that is when the body's immune system recognizes a part of the body as foreign and injures or goes to war against it. Rheumatoid arthritis and lupus are examples of autoimmune disorders.

Symptoms

PBC occurs in both men and women, but women get the disease 10 times more often than men. It usually begins between the ages of 30 and 60. Early in the disease, many patients have no symptoms. The only findings may be abnormal blood laboratory results. For example, a high level of the liver enzyme called alkaline phosphatase may be found in the blood. Itching and fatigue are common symptoms later in the disease. Itching is caused by bile entering the bloodstream.

As PBC progresses, other symptoms occur. There may be jaundice (yellowing of skin and eyes from excess bile in the blood), cholesterol deposits in the skin, fluid accumulation or edema, and darkening of the skin. Other immune related

MICROSCOPIC APPEARANCE OF LIVER



problems may also be present. For example, the tear and salivary glands may not function properly, causing dry eyes and mouth. Arthritis and thyroid problems may be present, and osteoporosis can develop in later stages. The bones become soft and fragile, leading to increased risk of fractures. The development of cirrhosis is the end result of PBC.

Diagnosis

PBC diagnosis is based on several pieces of information. Itching and fatigue alert the physician that bile ducts may be damaged. As previously mentioned, high levels of certain liver enzymes in the blood are important clues. Probably the most important laboratory test is one for mitochondrial antibodies. Mitochondria are the energy sources within cells. For unknown reasons a protein antibody develops against them in 95 percent of PBC cases. The physician must look at the whole picture to make the diagnosis of PBC.

Often the physician x-rays the bile ducts to rule out other causes of obstruction. This x-ray, called an ERCP, is performed under light sedation. A lighted, flexible endoscope is inserted through the mouth, stomach, and then